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A Discussion of the Limitations of Standardized Testing in a Pediatric Patient with Neuronal Migration Disorder: A Case Report

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Abstract

Background: Standardized, norm-referenced tests, such as The Bruininks-Oseretsky Test of Motor Proficiency, 2nd edition (BOT-2) and the Pediatric Balance Scale (PBS) are commonly used examination measures to assess progress of gross motor skills in pediatric patients. However, there is limited evidence to support the use of these measures in patients with chronic neurological conditions. **Case Description**: The patient is an 8-year-old girl. She has a medical diagnosis of Unilateral (left) Perisylvian Syndrome, a variant of neuronal migration disorder. The malformation of the left hemisphere has contributed to the following impairments: cognitive, motor, sensory and the gastrointestinal system, as well as occurrences of petit mal and atonic drop seizures. The patient has demonstrated minimal progressions, as well as regressions, of certain gross motor skills based on standardized testing. **Outcome Measures**: The main outcome measures utilized to determine patient progress were the BOT-2 and the PBS. A parent reported outcome measure, the Pediatrics Quality of Life Inventory Short Form (PedsQL 4.0 SF15) and parent interviews were also used to determine quality of life and participation levels of the patient. **Discussion & Outcome Considerations**: The purpose of this case report is to consider the discrepancies of standardized testing of pediatric patients with neurological disorders when compared to observed goal attainment in physical therapy, as well as parent reported participation and quality of life measures.

Keywords: Pediatrics; Neuronal Migration Disorder; Standardized Testing; Bruininks-Oseretsky Test of Motor Proficiency, 2nd edition (BOT-2); Pediatric Balance Scale (PBS); Pediatrics Quality of Life Inventory Short Form (PedsQL 4.0 SF15); Physical Therapy; Rehabilitation

Introduction

Pediatric physical therapy is a service provided to children and their families to assist each child in reaching their fullest potential to promote active participation in the home, school and community. The focus of physical therapy interventions is to address limitations in gross motor skill development and body function. A common way to measure a patient's progress in acquisition of gross motor skill is through the use of standardized norm-referenced measures of gross motor function, which became increasingly available during the 1970's and 1980's.[5] The process to identify developmental delays or disorders of motor development include developmental screening, examination and re-examination. Throughout the examination process, various measures are used, including discriminative and evaluative measures.[12] The BOT-2 is the most commonly used measure to evaluate motor deficits in children and adolescents with disabilities such as cerebral palsy, mild to moderate mental retardation, developmental coordination disorder, attention deficit hyperactivity disorder, and autism.[14] This standardized measure has provided clinicians with a valid measure reflective of typical childhood motor activities, which has demonstrated moderate to strong inter-rater and test-retest reliabilities.[8] However, this test is limited in its test-retest reliability for motor area composites for certain age groups. Therefore, clinicians should be familiar with these limitations when selecting this standardized test for the patient. In order to monitor the effectiveness of our chosen therapeutic interventions, it is critical to utilize reliable and sensitive measures that provide consistent results over time and are able to detect subtle changes in motor function.[14] This is a crucial element of the examination process and should be fit to the specific characteristics of the child.

The BOT-2, as well as the Pediatric Balance Scale, have shown to be reliable measures to determine delays in gross motor function across a variety of ages and diagnoses. In a review article of the BOT-2, researchers examined the tests ability to differentiate between clinical and non-clinical groups. They compared typical developing children to three clinical groups: children with developmental coordination disorder, children with mild to moderate mental retardation and children with high-functioning autism or Asperger's disorders. Each clinical group had significantly lower scores than its non-clinical comparison groups, supporting its ability to distinguish typical versus atypical populations.[8] When doing a literature search for sensitivity of specific populations with the use of BOT-2 and pediatric balance scale, there was moderate research surrounding the validity and reliability of populations such as cerebral palsy, developmental coordination disorder and autism spectrum disorder. However, many children seen by pediatric physical therapists do not fall under one of these three diagnoses, which have been researched at this time. As a result, clinicians may be utilizing ineffective standardized tests and measures to document progress of certain pediatric populations, for example children diagnosed with more severe or chronic neurological diagnoses. This could be negatively influencing a clinician's ability to detect subtle changes in progression of gross motor skills with the use of standardized test scores due to the limited evidence available to support the accuracy and validity of the BOT-2, as well as other supported tests and measures. Therefore, it is essential for additional research to be perused to expand our knowledge on the reliability and limitations of standardized testing of pediatric diagnoses which gross motor function is impaired. Thus, the purpose of this case report is to consider the discrepancies of standardized testing of pediatric patients with neurological disorders when compared to observed goal attainment in physical therapy, as well as parent reported outcome measures.

Case Description

The patient is an 8-year-old female, who has a recent formalized medical diagnosis of left (unilateral) Perisylvian Syndrome, a form of neuronal migration disorder. Neuronal migration is one of eight stages within the embryological neural development. These stages do not proceed linearly, but rather overlap as the brain and nervous system develops and matures.[10] The stage of development that is disrupted will influence the severity, the types of impairments and the child's prognosis for the future. Neuronal migration is when the post-mitotic neurons travel from the subventricular zone to their proper positions in order to create proper neural circuits.[9] However, when signals are absent or incorrect this can result in structurally abnormal or missing areas of the brain located in the cerebral hemispheres,

cerebellum, brainstem and/or hippocampus resulting in neuronal migration disorders. There have been reports of greater than 25 syndromes resulting from abnormal migration.[9] Some of these include schizencephaly, lissencephaly, pachygyria, neuronal heterotopias and polymircorgyria, the diagnosis of our patient described today. Polymicrogyria is when neurons move out to the cortical surface but organize abnormally to produce multiple small gyri with shallow sulci, which may be restricted to focal regions or diffuse across entire cortex.[11] There is a wide spectrum of severity, ranging from isolated grey matter nodules in the marginal zone or pier-ventricular region to board 'band' heterotopias in the sub-cortical white matter.[4] Our patient's formal diagnosis of Unilateral Perisylvian Syndrome has distinctive clinical and imaging features, which include a vertically oriented sylvian fissure continuous with the central/postcentral sulcus and includes variable polymicrogyric cortex features.[1] These features are consistent with recent MR imaging of our patient. Impressions from the MRI indicated extensive polymicrogyria tracking at left sylvian fissure, see Figure 1. The PET imaging correlated with the site of the gyral abnormalities, indicting asymmetric imaging with increased FDG uptake in the left frontotemporal and parietal lobe.

In a review of the patient's relative medical history, the patient did not have any significant pregnancy or birthing history reported by the mother at the child's physical therapy evaluation. However, at 3 months the patient was brought to the emergency department due to approximately one inch of swelling at the anterior fontanel, which prompted a CT scan, an MRI and a spinal tap. With the information gathered from these tests, she was diagnosed with a cortical brain malformation of the left hemisphere, as well as hemiparesis with right-sided weakness. Her mother indicated at the initial evaluation that the malformation of the left hemisphere has contributed to cognitive impairments, motor skill acquisition, sensory processing and disruption of the GI system. It is important to recognize that with this diagnosis, patient reported symptoms are variable based on the abnormality. However, there are often common features present including poor muscle tone and motor function, seizures, developmental delays, impaired cognitive development, failure to grow and thrive, difficulties with feeding, swelling in the extremities and smaller than normal head.[9] Perisylvian Syndrome specifically can present with contralateral hemiparesis/hemiplegia, which is consistent with our patient. It is also common for patients with a diagnosis of polymicrogyria to suffer from seizure disorders such as focal motor seizures, atypical absence seizures, atontic drop attacks and tonic-clonic siezures.[11] At 13-months, our patient had her first seizure and was diagnosed with focal epilepsy, experiencing petit mal seizures and atonic seizures localized to the left parietal lobe and prefrontal cortex. Over the last several months, parents have reported an increase in the frequency of seizure activity, which contributes to our patient's gross motor performance. It has been well supported that children with a medical diagnosis of epilepsy, like this patient, have been found to have significant dysfunction in gross motor and fine motor function.[2] With the most obvious deviations from their age-matched peers to be reported in running speed, balance, response speed and bilateral coordination.[3] It is important to recognize these are specific domains evaluated with the use of the BOT-2, as well as the Pediatric Balance Scale.

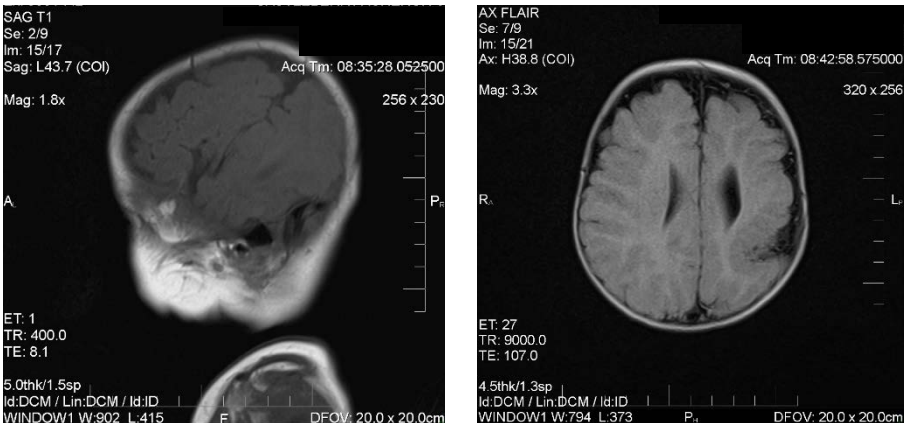

Figure 1. MRI sagittal and axial views; impressions indicative of features consistent with Unilateral Perisylvian Syndrome and extensive polymicrogyria tracking at left sylvian fissure.

The patient has been receiving developmental services, including physical therapy, occupational therapy and speech, which began at 7 months old due to delays in gross motor development (Table 1.). She continued services until the age of 2.5 years and discontinued services once developmental milestones were met. The family then returned to outpatient therapy services following a referral from her primary care physician at the age of 5 years-old due to established medical diagnosis of hemiplegia affecting the right side of the body, poor balance and generalized weakness. At the time of her evaluation, her mother reports falls were the primary concern to address with physical therapy services due to poor balance. The patient has received regular therapy services since. This allowed us to have access to formalized testing scores, monitoring her progression of gross motor performance and function over time. With her data, which will be presented in the following section, there are concerns of possible regression with the use of the BOT-2, as well as the pediatric balance scale. Therefore, this raises the question of patient progress and the factors that may be contributing to these findings and if standardized testing, such as the BOT-2, is appropriate for children with significant neurological impairments, such as this patient with a medical diagnosis of neuronal migration disorder.

Table 1. Typical Development compared to parent reported milestones for this patient.

Gross Motor Functions	Typical Milestones	Patient Reported Milestones
Rolling	4 months	6 months
Crawling	8 months	9 months *
Walking	12 months	16 months

**Parent reports patient did not complete true crawl [army crawl] before progressing to walking.*

Outcome Measures

The main outcome measures utilized to examine progress of our patient's gross motor function were the BOT-2 and the PBS. These are two well established test and measures examining activity limitations and body function limitations. We also completed the PedsQL 4.0 SF15 to determine quality of life and participation levels of the patient. Each of these outcome measures are supported by the Academy of Pediatric Physical Therapy.

Bruininks-Oseretsky Test of Motor Proficiency, 2nd edition (BOT-2)

The BOT-2 is a standardized measure of fine and gross motor skills of children and youth, ages 4 to 21 years of age. It is an evaluative measure used to characterize motor performance, intended for use by practitioners such as physical and occupational therapists, as well as researchers.[8] It is made up of a total of four motor area composites: fine manual control, manual coordination, body coordination, and strength and agility. As physical therapists we are interested in the testing of body coordination, which encompasses control and coordination of the large muscle groups used to maintain posture and balance. It assesses 15 total activities, such as jumping in place, tapping feet and fingers, standing heel-toe on balance beam, within two subcategories (bilateral coordination and balance).[8] The other motor area composite score important to note is strength and agility, which encompass aspects of fitness and coordination involved in casual play, competitive sports and other physical activities via 10 total activities.[8] The patient's raw scores are then calculated to z-scores and assigned a description category related to their age-equivalent peers. Finally, it is also important to recognize that the test–retest reliability and internal consistency of the total scale were excellent, and responsiveness was also found to be acceptable for all BOT-2 measures except the balance subtest.[14]

Our patient completed the BOT-2 standardized testing on three separates occasions over the course of approximately a year and a half. She completed testing at her chronological ages of 7 years, 3 months (Table 1), 7 years, 7 months (Table 2), and again at the age of 8 years, 10 months (Table 3). Although the BOT-2 has demonstrated good intra-tester reliability, it is important to note that our data in Table 1 was completed at a different facility following a move by the patient's family. This may contribute to some of the discrepancies in scoring, as well as patient performance working with a new therapist for the first time.

Table 2. BOT-2 scores completed approximately four months prior to initial evaluation at current outpatient therapy service clinic, age: 7 years, 3 months

	Total Point Scores	Scale (Standard) Scores	Z-Scores	Description Category
Body Coordination		17 (35)	-1.5	BA
Bilateral Coordination	12	10	-1.0	BA
Balance	22	7	-1.6	BA
Strength & Agility		15 (35)	-1.5	BA
Running Speed and Agility	11	4	-2.2	WBA
Strength (pushup – knees)	14	11	-0.8	A

Table 3. BOT-2 scores completed at initial evaluation for new location for outpatient therapy services, age: 7 years, 7 months

	Total Point Scores	Scale (Standard) Scores	Z-Scores	Description Category
Body Coordination		9 (27)	-2.0	WBA
Bilateral Coordination	6	5	-2.2	WBA
Balance	14	4	-2.3	WBA
Strength & Agility		8 (25)	-2.5	WBA
Running Speed and Agility	5	2	-2.6	WBA
Strength (pushup – knees)	7	6	-1.8	BA

Table 4. BOT-2 scores completed approximately 15 months after the initial evaluation, age: 8 years, 10 months.

	Total Point Scores	Scale (Standard) Scores	Z-Scores	Description Category
Body Coordination		5 (22)	-2.8	WBA
Bilateral Coordination	2	2	-2.6	WBA
Balance	13	3	-2.4	WBA
Strength & Agility		7 (23)	-2.8	WBA
Running Speed and Agility	7	3	-2.4	WBA
Strength (pushup – knees)	6	4	-2.2	WBA

Pediatric Balance Scale

The Pediatric Balance Scale is a modification of Berg's Balance Scale, developed as a tool to assess balance for school-aged children with mild to moderate motor impairments. It is a 14-item reference measure that examines functional balance which a child needs to perform in order to safely and independently navigate their home, school and community.[6] Functional balance is defined as the ability of a child to attain and maintain upright control during typical childhood activities of daily living, school and play.[5] Each item is scored on a scale from 0 (lowest function) to 4 (highest function). The score assigned is determined by the child's ability to perform the activity. The PBS has demonstrated good test-retest reliability and excellent inter-rater reliability when used with school-aged children with mild to moderate motor impairments.[6] There is also no significant difference among rating by different physical therapists for total test score.[5] However, similar to above when comparing scores of the BOT-2, we must consider the personal and environmental factors that may be contributing to our patients performance when comparing the two scores.

Our patient completed the pediatric balance test at a chronological age of 7 years, 3 months at her previous outpatient therapy location and for a second time at her chronological age of 8 years, 10 months at her current physical therapy location. These two tests were completed approximately a year and half apart. Our outcomes indicate this child is at a low risk of falls at this time. However, it is important to note that scores reported were completed with orthotics on and this may have improved her stability during testing. In future testing using the PBS, with this child, the assessing therapist should complete testing both with and without use of AFO's in order to determine her overall safety.

Table 5. Pediatric Balance Scale comparison; testing completed with use of bilateral AFOs.

Patient's Age	7 yrs, 3 mos	8 yrs, 10 mos
Total Test Score	53/56	45/56

Participation Measure

Participation and quality life outcome measures are valuable tools to indicate an individual's perceived physical and mental health over time. The Pediatric Quality of Life Inventory (PedsQL) is a parent and patient questionnaire that is ideal for assessing the impact of an intervention on health-related quality of life (HRQOL) over time. The PedsQL is a generic, multidimensional measure in children and adolescents, originally a 23-item questionnaire, for ages 2 to 18, covering the core dimension of health which include physical, social, emotional and role (school) functioning. This questionnaire includes self-reported children's versions, as well as the option for separate parent proxy reports.[13] An advantage of the PedsQL is the ability to create disease-specific modules to provide further insight into specific issues that impact HRQOL. For this parent reported outcome, we utilized the PedsQL 4.0 SF-15 (age 8-12), with a higher scaled score indicating better HRQOL. Our patient's mother filled out the questionnaire with a total score of 46.67. It is important to note that there is not a well-established standard for assessing pediatric health status. One study examined pediatric HRQOL using the PedQL 4.0, which indicated that for children older than 8 years old, the study recommended cutoff scores of 78 for total functioning.[7] From this information we can extrapolate that our patient's HRQOL is below a child's average score. This outcome measure will allow us as therapists to monitor this data and compare outcomes over time. In the future we could also include disease specific modules, such as the epilepsy module, to provide a more holistic picture of this child.

Table 6. PedsQL 4.0 Short Form, ages 8-12 parent reported outcomes

Domain	Raw Score	Mean Score
Physical Functioning	100/500	20.0
Emotional Functioning	300/400	75.0
Social Functioning	225/300	75.0
School Functioning	75/300	25.0
Total Score	700/1500	46.67

Discussion & Outcome Considerations

Based on completion of standardized testing with our patient, it was evident that compared to age-matched peers, she was well below average for her gross motor skill development. When analyzing her outcomes for the BOT-2 and the PBS over time, it appears that she has made little progress in the acquisition of her gross motor skills and functional balance. There are also concern that in certain areas that she may have even regressed. However, there are a few considerations that should be addressed when interpreting these scores. We also need to take her participation and quality of life measure into consideration, as well as her goal attainment in physical therapy following consistent therapeutic interventions over this time period.

The BOT-2 data points from the three standardized tests of this patient spans the course of approximately a year and a half, which we would expect to see improvements. However, there are considerations which need to be included from the data we have obtained. The first being that as a child's chronological age increases, they are compared to peers of the same age. Therefore, in the most recent standardized testing of the BOT-2 (Table 4), the patient increased in age level when

compared to a norm-referenced scores, which may have impacted the decrease in the z-scores and the descriptive category, from Table 3 to Table 4. However, when we look at the raw scores, especially for strength, speed and agility, and balance there is minimal change between the patient's performance. Some critics of standardized testing suggest that raw point scores are able to provide a more objective measure of evaluation than standard, age-related scores. This was based on the examination of individual children over time to specify improvement in their motor development, similar to our patient.[15] Therefore, with certain pediatric diagnoses, it may be a more accurate measure of a child's gross motor skills and allow us as clinicians to utilize sound clinical reasoning of a child's performance.

Environmental and personal factors have also been shown to play a role in the child's performance when completing standardized testing. Therefore, we must consider the change in environment faced by our child between the first test (Table 2) and second test (Table 3). Our patient's family had recently moved to a new state, as well as the change of working with a new therapist. Her mother reports that changes in schedule and environment increase the stress on her child and can lead to an increase in seizure activity, as well as an increase in poor behaviors (i.e. listening skills, impulsivity, etc.) Secondly, it is important to recognize how our patient's personal factors may be contributing to her performance. Thus, during the third and most recent standardized testing, the patient had been experiencing an increase in weekly seizure activity, as well as the return of atonic seizures, which had previously been well managed by medication and implantation of a Vagus Nerve Stimulator. These are critical factors that may have contributed to the fluctuations in our patient's performance over the course of 3 standardized tests. Similar to the BOT-2, her mother had reported prior to PBS testing, the patient had experienced atonic drop seizure the day before, which impacts her performance of gross motor skills the following day. As we have eluded to earlier, seizure activity in general has been linked to significant dysfunction in gross motor skills, specifically running speed, balance, response speed and bilateral coordination.[3] Therefore, this may have contributed to her decrease in scores over the course of a year and a half.

An additional consideration to reflect upon is the participation outcome measures, as well as goal attainment with physical therapy. Participation outcome measures are a subjective questionnaire of the patient's and parent's perspective of their physical and psychological functioning in various domains of a patient's life. They are a good tool to measure patient's progress and perception of their abilities over time. This will continue to be a beneficial tool for this child moving forward. We are also able to measure her improvements via patient goal attainment over the course of the year at her current outpatient therapy location. Throughout the patient's time working with physical therapy she has demonstrated improvement in her gross motor skills, which been recognized by her family. Some of the goals she has attained or that she is progressing toward include single limb balance, balance reactions with reduction of level of assistance provided, less compensatory movement patterns during the completion of core strengthening activities (i.e. sit-ups), and coordination of reciprocal stepping patterns on a balance bike. There is also parent report of improvements in daily life. Her mother shared some of these areas of improvement, which include significant improvements in her ability to control her body in space leading to the ability to complete stairs independently with less frequent falls. This has also increased her freedom during recess and the distance allowed away from her PARA, which has allowed for more natural interactions and play time with her peers. As a parent, she puts their family's experience into perspective stating that physical therapy has had a significant impact on her daughter's life. At the age of 7 months doctors told the family they didn't believe this patient would ever walk. Eight years later, although her skill level is similar to a 4-year-old, it is much better than her original prognosis provided by medical professionals.

Therefore, as the pressure from insurance companies and employers increase for clinicians to use validated standardized tests to evaluate patient progress, it is important to recognize when limitations are present with certain pediatric populations, like this patient. We need to use our clinical judgement to assess a patient's true performance based on a child's prognosis and diagnosis. Thus, when reflecting on this patient case report, it is evident that future research is necessary when examining the validity, responsiveness and sensitivity of standardized tests for more complex, specialized patient populations

in order to provide an accurate representation of patient progress. As clinicians, it is also important for us to consider the use of raw scores when using standardized and norm-referenced tests, as this may be a more effective tool in order to evaluate performance of gross motor skills over time, as skill acquisition may be subtle compared to their typically developing peers. Finally, it is always important to consider personal and environmental factors that may be influencing patient's performance. Therefore, use of participation and quality of life outcomes may be an effective tool to understand patient performance in their home environments and utilize this information in combination with parent reports of progress. This information will provide us with a holistic view of our patient and will allow us to recognize the patient's abilities and areas of concerns, allowing our therapeutic interventions to truly make a significant impact in our patient's day-to-day lives.

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